Anetoderma Secondary to Mid-dermal Elastolysis

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Abstract

Anetoderma usually presents as circumscribed, 1 cm to 2 cm patches and plaques of flaccid skin secondary to loss of dermal elastic tissue. Lesions often occur in the neck, upper extremities, chest, and back. On histopathology, one sees complete loss of dermal elastin involving the papillary and reticular dermis, with infiltration of plasma cells and histiocytes. A 40-year-old female with no significant medical history presented with multiple round, 1 cm to 2 cm lesions scattered on her upper back and chest. Skin biopsy demonstrated elastic-fiber loss localized to the mid-dermis along with a lymphohistiocytic infiltrate with elastophagocytosis and active inflammatory phase in the papillary and mid-dermal dermis. The histopathological findings were consistent with mid-dermal elastolysis with advancing inflammation, and the clinical features were consistent with anetoderma. The microscopic examination revealed an active inflammatory phase of mid-dermal elastolysis, supporting the postulated theory that MDE may be part of a continuous spectrum with anetoderma.

Case Report

A 40-year-old female with no significant medical history presented with multiple round, 1 cm to 2 cm lesions scattered throughout the upper back and chest. The lesions were characterized by lax, wrinkled skin with underlying palpable depression (Figure 1). They were often preceded by two to six months of local erythema and had increased in number over the past two years. No response was seen after topical steroids. Skin biopsy and elastic-fiber staining demonstrated elastic-fiber loss in the mid-dermis along with a lymphohistiocytic infiltrate with evidence of elastic-fiber phagocytosis and an active inflammatory background in the papillary and reticular dermis (Figures 2, 3 and 4). This is a case demonstrating the development of anetoderma as seen by the progressive inflammation and elastophagocytosis in the papillary and reticular dermis that developed in the setting of mid-dermal elastolysis (MDE).

Discussion

Mid-dermal elastolysis (MDE) is a rare acquired disorder of elastic-tissue degradation limited to the mid-dermis. It consists of a clear band of mid-dermal elastic-tissue loss as a result of inflammatory destruction of dermal elastic fibers. The elastic-tissue loss occurs as a result of inflammatory destruction of dermal elastic fibers. Remnants of abnormal elastic tissue and granuloma formation may be present, along with evidence of elastophagocytosis. Elastic tissue is usually preserved around hair follicles, resulting in perifollicular papules on the affected skin.
The condition was first described in 1977 by Shelley and Wood, and since then, there have been approximately 80 cases reported in the literature. It has a female predominance and presents clinically as diffuse, fine wrinkling on the neck, arms, and trunk in patients between the ages of 30 and 50 years.

It is classified into three types: type I, or classic type, with well-demarcated patches with wrinkling; type II, with peril follicular papular protrusions; and type III, with reticular annular patches with wrinkling.

Mid-dermal elastolysis has been reported to originate from involving sites of granuloma annulare that started as a patch, slightly indurated and violaceous eruption involving the neck and trunk and eventually became atrophic, pale and wrinkled. Urticaria, atopic dermatitis, Sweet's syndrome, phototoxic dermatitis, and pityriasis rosea have also been described to precede MDE.

Mid-dermal elastolysis can also occur in areas of preexisting erythema, which is consistent with our patient's clinical presentation. Anetoderma, also known as dermatitis maculosa atrophicans, is an elastic-tissue disorder that shows focal loss of elastic fibers in the dermis. The term "anetoderma" is derived from the Greek words "anetos," meaning slack, and "derma," meaning skin. Sac-like tumors that herniate upon palpation were first described by Schweninger and Buzzi in 1891, but anetoderma was first officially described in 1892 by Jadassohn.

It usually presents as multiple, circumscribed, 5 mm to 25 mm areas of flaccid skin with fine wrinkling that can occur in the neck, upper extremities, chest, and back. The lesions are skin color but can present with a blue-white discoloration. Affected areas of skin can herniate after palpation ("buttonhole" sign) and can show central depressions.

Anetoderma is classified as either primary (idiopathic) or secondary. Primary anetoderma is divided in two types: Jadassohn-Pelizzari type, which has preceding inflammatory lesions, and Schweninger-Buzzi type, which has no preceding inflammation. It is seen more commonly in women, and men usually occurs in individuals between 15 and 25 years of age. Secondary anetoderma may be associated with tumors, depositions, autoimmune disorders, infections, drugs and inflammatory cutaneous disorders. The loss of elastic tissue is usually localized to those sites of previous skin lesions caused by the primary disease.

On histopathology, there is focal loss of elastic fibers in the dermis with infiltration of plasma cells and histiocytes. The elastic fibers have an irregular shape and may be fragmented or engulfed by macrophages. Remnants of abnormal elastic tissue and granuloma formation may be present.

Activated macrophages and fibroblasts from existing inflammatory processes can destroy dermal stromal elements by releasing proteolytic enzymes. UV light exposure and autoimmunity against elastic fibers are thought to contribute to the condition. Defects in the synthesis of elastin and dysfunction of elastic-fiber digestion by metalloproteinases (MMPs) also seem to play a role. Giant cells and elastophagocytosis may be present in both mid-dermal elastolysis and anetoderma. Emer et al. described a case of anetoderma that was instigated by penicillin G to treat syphilis in an HIV-positive patient without any previous skin complaints.

Mid-dermal elastolysis and anetoderma, both disease entities resulting from elastic-fiber degradation, are differentiated histopathologically by the extent and location of elastic-fiber loss. The former consists of elastic-tissue loss localized to the mid-dermis, and the latter is characterized by elastic-fiber loss in the entire dermis.

It has been speculated that MDE and anetoderma are within the same spectrum of disorders, as they present similar histopathological configurations to different extents, suggesting MDE may evolve into anetoderma secondary to long-standing inflammation. Patient demonstrated a classic histopathology of MDE with secondary anetoderma resulting from an active inflammation and elastophagocytosis, lymphocytes, plasma cells and histiocytes extending to the papillary and reticular dermis.

Mid-dermal elastolysis and anetoderma need to be differentiated from other connective-tissue diseases affecting elastic fibers, including cutis laxa, pseudoxanthoma elasticum (PXE), and PXE-like papillary dermal elastolysis. Post-traumatic scars, peril follicular elastolysis, papular elastorrhexis, pseudoxanthoma elasticum, focal dermal hypoplasia (Goltz syndrome), and nevus lipomatosus are other entities that may be included in the differential diagnosis of anetoderma.

Cutis laxa is an entity with redundant and loose skin seen on the eyelids, cheeks, shoulder girdle, abdomen and neck. It presents clinically with premature aging secondary to loose skin folds with or without internal organ involvement. In this condition, the whole dermis is affected with diminished and fragmented elastic fibers, and it can occur in an acquired or hereditary form.

Pseudoxanthoma elasticum (PXE) and PXE-like papillary dermal elastolysis present as cobblestoning yellow papules and redundant folds in flexor areas. The former can be associated with oculocutaneous involvement. It occurs in sites of previous scars, axilla, groin, and lateral neck and consists of clumped and calcified elastic fibers in the mid-dermis. The latter is seen in inflammatory folds, lower abdomen, axilla, and neck, and has a band-like pattern of clumping and fragmentation of elastic tissue in the papillary dermis.

Intralesional triamcinolone injections and systemic administrations of dapsone, aspirin, penicillin G, vitamin E, and inositol niacinate have been used to treat existing anetodermal lesions with success. Administration of hydroxychloroquine, colchicine, and aminocaproic acid, as well as surgical excision, have resulted in some improvement. Cho et al. reported anetoderma presenting after Stevens-Johnson syndrome successfully treated with ablative carbon dioxide fractional laser. Lasers destroy the hydrogen bonds in the collagen triple helix, instigating an inflammatory cascade that is believed to be responsible for rebuilding stable and more native-like collagen and elastic fibers, thereby reverting the pathological process.

Conclusion

We report a case demonstrating the development of anetoderma as seen by progressive inflammation and elastophagocytosis in the papillary and reticular dermis that developed in the setting of mid-dermal elastolysis (MDE). The histopathological findings were consistent with mid-dermal elastolysis with active inflammation, and the clinical features were consistent with anetoderma. The histopathological examination revealed elastic fiber loss localized to the mid-dermis along with a lymphohistiocytic infiltrate with elastophagocytosis and active inflammatory phase in the papillary and mid-recticular dermis. Such areas of active inflammation indicate the development of anetoderma in the pre-existing MDE background as elastic-fiber degradation spread beyond the mid-dermis to involve the papillary and reticular dermis.

Anetoderma often occurs from existing inflammatory processes that lead to elastic-fiber destruction, thus the overall findings in this case may support the theory that MDE is part of a continuous spectrum with anetoderma.

References

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